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Management of Vascular Malformations

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Background: Even though vascular malformations are well categorized, further details are relatively unknown. Of treated patients regarding the frequency, demographic distributions, and other related factors by multivariate regression analyses in proportion to total vascular malformations, methods of treatment and how to manage them have not been elucidated thoroughly.
Methods: From January 2006 to March 2012, consecutively treated patients with vascular anomalies were included in this investigation at least 1-year follow-up.
Results: Of the total of 123 cases, 86 females and 37 males, the mean follow-up was 3.5±1.68 years, and the frequency of treatment was 1–8 times (1.8±1.30). Surgery was performed for 22 cases (17.9%) of venous malformations and arteriovenous malformations. In multivariate regression, the frequency of treatment was significantly correlated with the length of follow-up ($P < 0.001$), age ($P < 0.05$), and type of malformations ($P < 0.05$) ($R^2 = 0.18$). Need for surgery was significantly increased with age at odds ratio (OR) of 1.06 [95% confidence interval (CI), 1.03–1.80] ($P < 0.001$), and head/face/neck, and upper limb are more performed at OR of 0.24 (95% CI, 0.07–0.85) ($P < 0.05$). The satisfaction score varied from 1 to 5 (3.9±0.68). Complications occurred in 3 cases (2.4%). In logistic regression of complications, the OR of the satisfaction score was 0.13 (95% CI, 0.02–0.80) ($P < 0.05$).
Conclusions: Treatment of vascular malformations is an integral part of multidisciplinary approaches. Venous malformations are more frequent in combination surgery, and if there are fewer complications, the patients' satisfaction increases. *(Plast Reconstr Surg Glob Open 2014;2:e128; doi: 10.1097/GOX.0000000000000079; Published online 27 March 2014.)*

Vascular malformations include capillary malformation (CM), venous malformation (VM), lymphatic malformation (LM), arteriovenous malformation (AVM), and their combinations,[1] and they are distinct from vascular tumors regarding clinical appearance, imaging, and histopathological characteristics. VM is the most common type of vascular malformation, and the overall incidence of VM is reportedly 65% of the total 1.5% of all congenital vascular malformations found in a general population.[2] AVMs occur with equal frequency in men and in women, with 40–60% of lesions apparent at birth and an additional 30% becoming obvious during childhood.[3] AVMs are categorized into 4 different stages proposed by Schobinger and accepted by the International Society for the Study of Vascular Anomalies.[4] The natural progression and recurrence of AVMs

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in children are likely to occur before adulthood and may exacerbate with resection either with or without embolization.[5] LMs are developmental anomalies of the lymphatic system that result in defects in lymphatic flow. Both macrocystic and microcystic LMs are often present at birth. Macrocystic LMs are more frequently located in the neck and axilla and are often referred to incorrectly as cystic hygroma,[6] whereas microcystic LMs become apparent after complications of infection or bleeding and are predominantly located in the proximal limbs, upper extremities, axillae, and the chest. CM is a slow-flow anomaly, occurs in 0.3% of infants, and is sometimes diagnosed as bruising or erythema from birth trauma. CM is a congenital anomaly that is believed to be a vasculature developmental error and can be clinically diagnosed. CM may be mimicked by cutaneous erythema overlying deeper AVMs. Doppler ultrasound, computed tomography, and magnetic resonance imaging (MRI) may be helpful in diagnosing arteriovenous fistula or shunting.[7] Despite the availability of embolo/sclerotherapy for vascular malformations,[8] the role of surgical removal and subsequent reconstruction is relatively low due to the risk of bleeding during surgery and inaccessibility for excision of the entire vascular malformation, which may result in deterioration of future treatment. In patients undergoing embolo/sclerotherapy, the incidence of soft-tissue injury and neuropathy was reported to be 11.9% and 8.6%, respectively, in 573 patients, and surgical treatment such as escharectomy, skin grafting, and amputation was employed for 41.2% of the soft-tissue injury cases to rescue and restore the defects.[9] We analyze all the treated patients for frequency, demographic distributions, and other related factors by multivariate regression analyses in proportion to total vascular malformations, and methods of treatment and management are discussed.

MATERIALS AND METHODS

From January 2006 to March 2012, consecutive treated patients were included in this investigation of vascular anomalies.

All the patients were followed for at least 1 year after the last treatment. The pretreatment before including in our study was divided into 4 subcategories: 0 = no previous treatment, 1 = 1- to 3-time treatment, 2 = 4- to 10-time treatment, and 3 = over 10-time treatment. The subjective satisfaction score (1, lowest to 5, highest) was collected at the last outpatient clinic for each patient. The agent used for embolization was mainly N-butyl cyanoacrylate, and for large and high-input lesions, a platinum-feathered coil was used, while absolute ethanol was used for sclerotherapy at 0.1 mL/injection under the echo guidance at maximum of 0.6 mL/body weight (kg). When embolization and sclerotherapy were combined, a 24- to 72-hour interval was employed. Surgery could be performed at the time of sclerotherapy.

When the lesion is so obviously small and well-defined, after sclerotherapy, surgically resection is followed. Or redundant tissue may be suspended after sclerotherapy. Surgical reconstruction was followed if the lesion is considered completely resected.

Clinical assessment and imaging examinations such as Doppler ultrasonography, computed tomography, MRI, and angiography preceded any treatment such as transcutaneous embolization and percutaneous sclerotherapy for AVMs or sclerotherapy alone for other vascular malformations. When surgery was implemented, all were followed by embolization and sclerotherapy (embolo/sclerotherapy) or sclerotherapy.

Patient Consent and Ethical Disclosure

All treatments in this clinical series were approved by the Internal Review Board of Nagasaki University (approved number 10032690), and informed consent forms are included in the internal review board format.

Statistics

Results are expressed as the mean ± SD. The multiple linear regression test and logistic regression test were used for multiple variable analysis. Statistical significance was defined as $P < 0.05$. All analyses were performed using IBM SPSS Statistics, version 21 (Japan IBM Co. Ltd, Tokyo, Japan).

RESULTS

All consecutive cases were 123: 86 female (69.9%) and 37 male (30.1%). There were 85 cases of VMs, 19 AVMs, 8 LMs, 4 CMs, 2 capillary and venous malformations (CVM), and 5 lymphatic and venous malformations (LVMs), and patients were treated in the Department of Plastic and Reconstructive Surgery, Nagasaki University Hospital. The distribution of anatomical locations was 59 cases (48.0%) in the head/face/neck, 18 cases (14.6%) in the upper limbs, 11 cases (8.9%) in the torso, 34 cases (27.6%) in the lower limbs, and 1 case (0.8%) in the head/face/neck and torso (Table 1).

Table 1. Follow-up, Frequency of Treatment, and Number of Surgeries

Type and Location	Follow-up (Y) (Mean ± SD)	Frequency of Pretreatment* (Mean ± SD)	Frequency of Treatment (Mean ± SD)	Surgery (Number)	
Type					
VM, $n = 85$	3.3±0.18	1.2±0.63	1.5±0.98	19	
AVM, $n = 19$	3.7±0.37	2.2±0.69	2.7±1.67	3	
LM, $n = 8$	4.7±0.44	1.9±0.64	2.3±1.58		
CM, $n = 4$	2.5±0.46	1.5±0.58	3.5±2.08		
CVM, $n = 2$	6.4 and 4.6	3 and 3	4 and 1		
LVM, $n = 5$	4.3±0.90	1.8±0.84	1.8±0.84		
Location				VM	AVM
Head/face/neck, $n = 59$	3.7±0.23	1.9±1.25	1.9±1.25	11	2
Upper limb, $n = 18$	3.5±0.35	1.3±0.46	1.3±0.46	2	
Torso, $n = 11$	3.3±0.42	1.5±0.69	1.5±0.69	2	
Lower limb, $n = 34$	3.3±0.30	2.1±1.71	2.1±1.71	3	1
Head/face/neck and torso, $n = 1$	1.1	1	1	1	

*Frequency of pretreatment is set: 0 = no previous treatment, 1 = 1- to 3-time treatment, 2 = 4- to 10-time treatment, and 3 = over 10-time treatment.

Systemic vascular malformations, such as Klippel-Trenaunay syndrome,[10] Parkes-Weber syndrome,[11] and Sturge-Weber syndrome,[12] were excluded from this study. The mean age was 27.6±21.16 (1–88 years old) for VMs, 40.9±19.89 (11–80 years old) for AVMs, 19.8±20.74 (2–61 years old) for LMs, 54.0±4.97 (49–60 years old) for LMs, 33.0±12.73 (24–42 years old) for CVMs, and 13.0±7.21 (6–24 years old) for LVMs. The mean follow-up was 3.5±1.68 years. Mean follow-up for VM, AVM, LM, CM, CVM, and LVM was 3.3±0.18 years (1–7.2), 3.7±0.37 years (1.2–6.7), 4.7±0.44 years (2.9–7.0), 2.5±0.46 years (1.2–3.3), 5.5±0.90 years (4.6–6.4), and 4.3±0.60 years (1.9–5.2), respectively. Mean follow-up for the head/face/neck, upper limbs, torso, lower limbs, and head/face/neck and torso was 3.7±0.23 years (1.0–7.0), 3.5±0.35 years (1.1–6.5), 3.3±0.42 years (1.2–5.2), 3.3±0.30 years (1.0–7.2), and 1.1 years, respectively.

The frequency of treatment was 1–8 times (1.8±1.30): 1.5±0.98 (1–6) times for VM, 2.7±1.67 (1–8) times for AVM, 2.3±1.58 (1–5) times for LM, 3.5±2.08 (1–6) times for CM, 2.5±2.12 (1–4) times for CVM, and 1.8±0.84 (1–3) times for LVM. The frequency of treatment was 1.9±1.25 (1–6) times for the head/face/neck, 1.3±0.46 (1–2) times for upper limbs, 1.5±0.69 (1–3) times for the torso, 2.1±1.71 (1–8) times for lower limbs, and once for the head/face/neck and torso.

Surgery was performed for 22 cases (17.9%), subdivided into 19 of 85 cases (22.4%) for VM and 3 of 19 cases (15.8%) for AVM. Thirteen of 59 cases (22.0%) in the head/face/neck, 2 of 18 cases (11.1%) in upper limbs, 2 of 11 cases (18.2%) in the torso, 4 of 34 cases (11.8%) in lower limbs, and 1 of 1 case (100%) in the head/face/neck and torso underwent surgery. In VMs, 11 of 19 cases (58.0%) were in the head/face/neck, 2 of 19 (10.5%) in upper limbs, 2 of 19 (10.5%) in the torso, 3 of 19 (15.8%) in lower limbs, and 1 of 19 (5.3%) in the head/face/neck and torso. In contrast, in AVMs, 2 of 3 (66.7%) cases were in the head/face/neck and 1 of 3 cases was in lower limbs (Table 1). Considered complete surgical resections were 14 cases and 1 case for AVM and 13 cases for VMs. An AVM case of the temporal head was reconstructed with free-vascularized anterolateral thigh flap. Nine cases of head/face/neck VMs are considered completely resected; 2 cases of upper limb VMs and 2 cases of torso VMs are considered completely resected. The mean follow-up for surgical cases was 3.4±0.46 (1.0–7.1) years, whereas 3.5±0.16 years for nonsurgical cases.

In multivariate regression analysis, when the frequency of treatment was set as a criterion variable and explanatory variables were defined for age, location, type, and sex, age ($P < 0.05$), type ($P < 0.05$), and follow-up ($P < 0.001$) demonstrated the significant difference ($R^2 = 0.18$, $F = 2$).

In multivariate regression analysis, when follow-up was set as a criterion variable and frequency and explanatory variables were defined for age, location, type, and sex, statistical significance was observed for frequency ($P < 0.001$) ($R^2 = 0.13$, $F = 2$).

In a multivariate regression analysis, when the pretreatment was set as a criterion variable and explanatory variables were defined for age, follow-up, location, type, frequency, and sex, statistical significance was observed in frequency ($P < 0.001$), location ($P < 0.05$), type ($P < 0.05$), and sex ($P < 0.02$) ($R^2 = 0.85$, $F = 2$). More pretreatment leads to more frequent interventions in this study, head/face/neck location demonstrates more significant than other locations, CVM and AVM are more than other types, and females are more frequent in pretreatment.

In multivariate logistic regression analysis, backward linear regression step-wise regression with or

without surgery, explanatory variables were set for follow-up, age, frequency of the treatment, location, type, and sex, and the odds ratios (OR) of age and type were 1.06 [95% confidence interval (CI), 1.03–1.80] ($P < 0.001$) and 0.24 (95% CI, 0.07–0.85) ($P < 0.05$), respectively. Surgery was only performed for VM and AVM and showed significantly greater frequency in VM compared with AVM (Table 2). More multidisciplinary treatment with embolo/sclerotherapy and surgery are utilized in AVM.

The satisfaction scores ranged from 1 to 5 (3.9±0.68): 3.9±0.07 (2–5) for VM, 3.6±0.19 (1–5) for AVM, 4.3±0.25 (3–5) for LM, 3.5±0.29 (3–4) for CM, 4 for CVM, and 4 for LVM. The satisfactory scores for location were 4.0±0.08 (2–5) for the head/face/neck, 3.6±0.18 (1–4) for upper limbs, 4.0±0.19 (3–5) for the torso, 3.8±0.11 (2–5) for lower limbs, and 3 for the head/face/neck and torso.

Complications occurred in 3 cases (2.4%), of which 1 case was in AVMs and showed tissue necrosis after sclerotherapy and 2 were in VMs, one of which demonstrated incomplete facial nerve paralysis and the other suffered massive bleeding and required a blood transfusion to recover, because systemic anticoagulation and antihemorrhage capacities were limited due to massive and multiple malformations. In multivariate logistic regression analysis, backward LR step-wise regression with or without complications, explanatory variables were set for follow-up, age, satisfactory score, frequency of the treatment, location, type, and sex, and the OR of the satisfaction score was 0.13 (95% CI, 0.02–0.80) ($P < 0.05$). When there were fewer complications, the satisfaction score increased.

DISCUSSION

Treatment of vascular malformations is an integral part of percutaneous embolization, percutaneous sclerotherapy, and surgery. Preoperative embolo/sclerotherapy is beneficial for decreasing intraoperative bleeding and defining lesion margins to solidify lesions, especially in slow-flow vascular malformations. In the treatment of patients with vascular malformations, in a single-center analysis of 1130 cases of VM and LVMs and 329 AVMs, surgery was performed for VMs and LVMs (5% with embolo/sclerotherapy and 4% without), while 15% underwent embolo/sclerotherapy for AVMs.[8] All of our cases in this study were preceded by presurgical embolo/sclerotherapy or sclerotherapy, with 22.4% surgeries for VMs and 15.8% for AVMs. Surgeries were performed only for VMs and AVMs because both VMs and AVMs are of concern regarding esthetics and restoring functions,[5,13] and AVM patients who are surgically resected with or without embolization exhibit less re-expansion than those treated with embolization alone, and recurrence was less probable when the AVM stage (Schobinger staging) was lower in multivariate logistic regression analysis.[5] Moreover, it may reflect that the cases are more defined and more easily accessible for surgical removal and reconstruction. Also, all the cases in this study underwent at least one treatment and the data settings may have been different. Therapeutic management of vascular malformations is an integral part. In our 123 series, only 5 cases are first time treatment cases and usually require multiple sessions and modalities. Previously treated cases sometime cause more complicated pathology as the presence of the scar tissue and altered microvasculature of the lesion. In VMs, surgical interventions are required for control of pain, bleeding, nerve impairment due to compression and extensively deterioration in function, and aesthetics. When the lesion is well surrounded, surgical resection and reconstruction is recommended.[14]

In our series, only 19 of 85 cases (22.4%) can lead to surgical resection and reconstruction, because the lesions are beyond the border of healthy deeper tissue and margin of the lesions is obscure to clarify, partly due to the previous treatment.

In AVMs, microvascular proliferation is correlated to the rate of the flow of the lesion regardless of the previous embolization[15]; thus, when the lesion seems active and expanding with fast flows, therapeutic interventions may be necessary to control of subsequent deterioration of the lesion or skeletal impacts to the pediatric patients.

In a follow-up of an average 3 years and 6 months, the mean frequency of treatment was 1.8 per case in our series. Surgery was performed in 17.9% of the cases in this study and according to logistic regression analysis. Complications occurred in 3 cases: 1 AVM and 2 VMs. In the AVM case, the complication was tissue necrosis due to fast flow leading to carriage of the injected absolute agent to peripheral sites; one massive facial VM resulted in an incomplete facial nerve, the buccal branch of the facial nerve, paralysis; and another VM case suffered continuous bleed-

Table 2. Multivariate Logistic Analyses of Variables to Predict Surgery

Variables	*P*	Odds Ratio	95% CI
Follow-up	0.657	0.93	0.68–1.28
Age	0.000*	1.06	1.03–1.08
Sex	0.079	2.78	0.89–8.72
Location	0.277	0.79	0.51–1.21
Type	0.027†	0.24	0.07–0.85
Frequency	0.647	1.11	0.71–1.74
Satisfactory factor	0.831	0.91	0.39–2.12

*$P < 0.001$.
†$P < 0.05$.

ing after sclerotherapy. Soft-tissue loss was observed in 68 of 573 cases (11.8%) and was greatest in AVMs, 42 of 143 cases (29.4%). Soft-tissue loss spontaneously recovered within 2.7 months, and the final rate was 28 of 573 cases (4.9%). Neuropathy was observed in 49 of 573 cases (8.6%), with VM demonstrating 30 of 273 cases (10.9%), and the final rate was 7 of 573 cases (1.2%) at 5.3 months.[9] Our patients were assessed at least 1 year after the last treatment, and 1 of 19 AVMs (5.3%) demonstrated skin loss, in which the lesion lay along the digital vessels and injected absolute ethanol ran distally to the finger-tip. The rate of neuropathy in VM was 1 of 85 cases (1.2%), which was a massive and enlarged lesion, the frequency was 6 times, and appearing facial nerve courses may have been masked, with both rates being very comparable. There were no complications related to surgery. When the patient's satisfaction was greater, the complication rate was inversely correlated, with an OR of 0.13. In a cross-sectional study of 158 VMs, female gender (OR, 4.49; 95% CI, 1.24–16.28), no or delayed visualization of drainage vein (OR, 9.22; 95% CI, 1.79–47.51), and a well-defined margin on MRI (OR, 13.38; 95% CI, 2.84–63.12) were independent predictors of good responders on multivariate analysis.[16]

In AVM, angiographic types and treatment are recommended according to different accessibility to the lesion, such as transarterial access, direct puncture, and transvenous access.[17]

CONCLUSIONS

Further objective analyses of the margin of lesions and drainage veins for VMs in surgery and appropriate combinations with different access and pretreated AVMs should be defined and appropriate follow-up and definitive removal of the lesion should be carefully determined in relation to the timing of the therapy and methods.

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Vascular Malformations That Were Diagnosed as or Accompanied by Malignant Tumors

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BACKGROUND Vascular malformations can mimic malignant tumors, and the coexistence of both types of lesions can limit and interfere with treatment. A consecutive series of vascular malformations that were treated and evaluated in a single institute and cases involving vascular anomalies combined with malignancies or malignancies that were treated as vascular anomalies were analyzed.

OBJECTIVE Absolute ethanol is used in the treatment and management of vascular malformations and is sometimes administered before a definitive diagnosis has been obtained, despite the fact that some vascular lesions are subsequently revealed to be malignant tumors. This study discusses such cases.

MATERIALS AND METHODS From January 2006 to August 2012, 139 patients were treated for vascular malformations at Nagasaki University Hospital and were followed up for a minimum of 1 year.

RESULTS Four malignant lesions coexisted with or were misdiagnosed as vascular malformations, including a malignant peripheral nerve sheath tumor located in the chest, a hemangiopericytoma of the palate, an adenoid cystic carcinoma of the cheek, and a squamous cell carcinoma of the cheek. Thus, malignant lesions were detected in 2.88% of cases in which vascular malformations were preoperatively diagnosed.

CONCLUSION When treating vascular malformations, it is advisable to be aware of the possibility of malignancy.

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When diagnosing vascular malformations and distinguishing them from tumors, especially malignant tumors, imaging using ultrasound (US), magnetic resonance imaging (MRI), and computed tomography (CT) can be used to clarify the characteristics and extent of the target lesion. Although arteriovenous malformations (AVM) are clinically easier to detect because they exhibit pulsation and niduses on MRI and CT and vascular malformations display evidence of shunt-flow within or around them on US, other types of lesions, including malignant tumors, can also display similar imaging features.

Once a diagnosis of vascular malformation has been made, embolization, or percutaneous sclerotherapy is used to control any bleeding, which enables resection and reconstruction in selected cases.[1–4] However, some vascular lesions coexist with malignant tumors or are treated as vascular lesions or tumors without a precise diagnosis being obtained. Hemangiopericytoma, a rare vascular tumor, exhibits marked enhancement together with blood flow and flow voids. In addition, hemangiopericytomas can arise in neurovascular bundles and can cause marked hemodynamic changes due to occluded sinuses and aberrant pericytes. Furthermore, cranial hemangiopericytomas can result in marked arteriovenous shunting.[5]

Thus, tumors can mimic vascular malformations during clinical and imaging examinations. As a result, malignant tumors that display pulsation, are well demarcated, and/or exhibit the typical imaging and

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clinical features of vascular malformations are sometimes misdiagnosed as vascular malformations. However, a previous report described the use of ethanol for US-guided endovascular sclerotherapy or fluoroscopy as a treatment for vascular malformations,[3] and the percutaneous US-guided injection of ethanol into hepatocellular carcinoma has also been demonstrated to regulate tumor volume and reduce the size of residual tumors[6] or recurrent malignant tumors[7]; therefore, treatments that would normally be applied to vascular malformations can also be used in cases that might involve malignancy.

In the salivary glands and neighboring regions, neoplasms are generally distinguishable from normal glandular tissue. In fact, a previous study found that 95% of major salivary gland lesions could be delineated and that it was possible to accurately identify intraglandular vessels within the parotid gland.[8] In addition, MRI is useful in assessing the perineural spread of malignant tumors. Among the malignant tumors that affect the salivary glands, adenocystic carcinoma shows the greatest propensity for perineural and perivascular spread. Both US and MRI have been demonstrated to be reliable for determining whether salivary gland tumors are benign or malignant.[8,9] Regarding malignant peripheral nerve sheath tumors (MPNST), it has been reported that arteriovenous fistulae can arise from the affected nerve in pediatric cases.[10] However, during US of MPNST, prominent intramass vessels that exhibit marked vibration and little resistance can mimic the findings of vascular malformations. On MRI scans obtained using the T2 window, MPNST exhibit high-intensity signals, but no target sign, and MR angiography has been demonstrated to be useful in depicting arteriovenous fistulae within MPNST. Neurofibromatosis Type 1 (NF-1) occurs in up to 50% of patients with MPNST; however, it has also been demonstrated in a young child with a lumbar epidural AVM.[11] A previous report described a case of squamous cell carcinoma (SCC) involving a coexisting massive lymphaticovenous malformation, in which it was necessary to excise the entire lymphaticovenous malformation to clear the patient's airway, and both the malformation and tumor were successfully removed.[12] Among vascular anomalies, which include both vascular malformations and hemangiomas,[13] the cytoplasmic endothelia of infantile and congenital hemangiomas were found to be immunohistochemically positive for Wilms tumor 1 (WT1) protein, whereas vascular malformations such as lymphatic and venous malformations did not exhibit WT1 positivity.[14] In a previous study of 117 vascular neoplasms and 50 vascular malformations, only the AVM exhibited positive immunostaining for WT1, and the majority of vascular malformations demonstrated no WT1 expression.[15] Furthermore, a mutation in the phosphatase and tensin (PTEN) homolog tumor-suppressor gene is present in 54% of vascular anomalies, 86% of which are fast-flowing vascular malformations.[16] Here, cases in which the similarities between certain tumors and vascular malformations led to misdiagnosis are discussed.

Patients and Methods

From January 2006 to August 2012, 139 patients were treated for vascular malformations at the Department of Plastic and Reconstructive Surgery, Nagasaki University Hospital. Among the vascular malformations, 93 (66.91%), 20 (14.39%), 12 (8.63%), 4 (2.88%), 3 (2.16%), 5 (3.60%), and 2 (1.44%) were arteriovenous, lymphatic, capillary, capillary–venous, lymphaticovenous, and capillary–lymphatic malformations, respectively. The mean age of the patients was 28.2 ± 22.13 (range, 1–88 years). All of the patients were of Asian origin. The patients were first clinically evaluated with Doppler US, followed by MRI and CT. Patients with AVM underwent angiography, and transarterial embolization was performed in some cases. Direct percutaneous sclerotherapy was performed with the assistance of duplex Doppler US using absolute ethanol at a dose of 0.1 mL per injection and a maximum volume of 0.5 mL × body weight (kg). For patients with superficial slow-flowing lesions, 3% polidocanol, which was prepared as a foam with a 1:4 polidocanol:air ratio in a syringe, was added and was administered at a maximum dose of 4 mL. Four patients who were initially diagnosed with vascular malformations were subsequently found to have low-grade to high-grade malignant tumors by histologic and immunohistochemical examinations. Thus, malignant lesions accounted for 2.88% of our series.

Patient Consent and Ethics Disclosure

All treatments in this clinical series were approved by the internal review board of Nagasaki University (IRB) (approval number 10032690), and informed consent forms were obtained in the format requested by the IRB.

Cases Involving Malignancy

Case 1

A lesion that was diagnosed as a slow-flow venous malformation, but was subsequently found to be a malignant peripheral nerve sheath tumor

A 39-year-old male patient was referred to us with a salmon pink, smooth, hard, and heterogeneously elastic mass measuring 5.5 × 4 cm in the anterior region of his chest. The mass had first been noticed by the patient 25 years ago (Figure 1). Subsequently, it gradually enlarged, and internal and external bleeding occurred on several occasions. Ultrasound revealed a relatively homogeneous hypoechoic lesion that had invaded the subcutaneous adipose tissue, and some blood flow was observed inside and around the mass. T2-weighted MRI images demonstrated a multilobular lesion that displayed high-intensity signals and had not invaded the surrounding muscle or bone tissue. Some regions that exhibited low signal intensity were considered to be phleboliths or thrombi. A blood vessel that connected the lesion to the internal mammary blood vessels was detected on MRI.

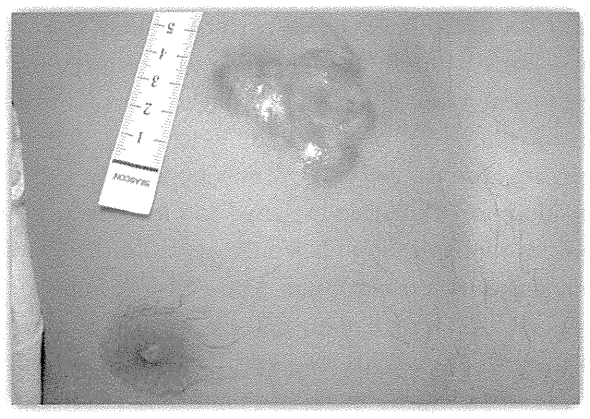

Figure 1. A 39-year-old male patient was referred with a salmon pink, smooth mass measuring 5.5 × 4 cm.

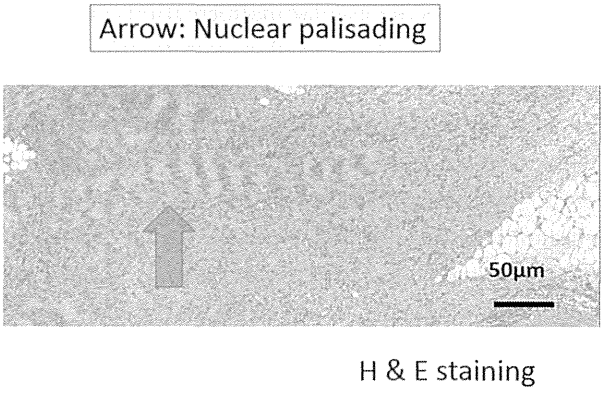

Figure 2. Histologic examination of the mass shown in Figure 1 (hematoxylin and eosin stain staining). The arrow indicates the observed storiform pattern and palisading nuclei. Magnification ×40.

Overall, the pretreatment data were compatible with a slow-flow vascular malformation. Thus, the lesion was subjected to echo-guided transcutaneous sclerotherapy using absolute alcohol, followed by excisional biopsy to histologically confirm the suspected diagnosis. The biopsy sample contained cells with a storiform pattern and some palisading nuclei (Figure 2). Positive immunohistochemical staining of S100 and the presence of interstitial edema led to a diagnosis of MPNST or malignant schwannoma (Figure 3). Three weeks after the microscopic examination, the tumor and a 3-cm margin were resected to the depth of the pectoralis muscles and sternal periosteum. The excision margins were examined on frozen sections and were confirmed to be negative, and a 6- × 18-cm ipsilateral

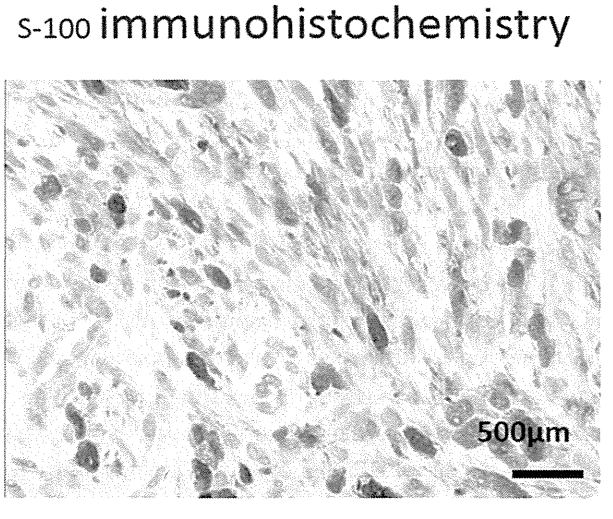

Figure 3. Immunohistochemical examination of S100 expression. The immunoreactive cells displayed a fusiform shape. Magnification ×400.

latissimus dorsi musculocutaneous flap was used to cover the defect. At 36 postoperative months, the patient's course was uneventful (Figure 4), and no clinical or positron emission tomography–CT findings that were indicative of recurrence or metastasis were observed.

Case 2

A lesion that was diagnosed as an arteriovenous malformation, but was subsequently found to be a hemangiopericytoma

A 26-year-old male patient had an elastic hard mass (diameter, 5 cm) with a smooth surface in his left palate, which has been present for the past 2 months, and the surface of the lesion exhibited recurrent erosion. However, the patient did not experience any acute pain or difficulty in swallowing (Figure 5). A multidetector CT scan demonstrated a vascular-rich lesion extending from the upper to middle mucous membranes of the larynx (Figure 6). In addition, an MR angiogram depicted abundant vascular structures in the surrounding regions, but not in the core part of the mass. Color Doppler detected mixed-type hypervascularity around the smooth mass. An AVM was initially suspected, but other vascular tumors, such as juvenile angiofibroma, were considered as possible diagnoses. The therapeutic plan involved transarterial embolization of the maxillary arteries and other branches of the external carotid artery, and after 24 hours, echo-guided transmucous membrane sclerotherapy was performed. The mass under the mucous membrane was enucleated, and the total blood loss during the procedure was 50 mL. There had been no recurrence at 4 postoperative months. A histologic examination demonstrated a stag-horn vascular pattern and slit-like vascular channels, and CD34 immunohistochemical staining revealed strong immunoreactivity, which was distinct from the CD34 staining patterns exhibited by solitary fibrous tumors,

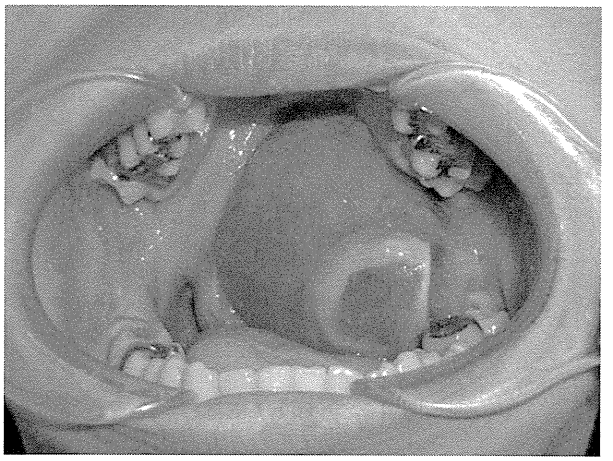

Figure 5. A 26-year-old male patient had noticed a hard elastic mass with a smooth surface (diameter, 5 cm) in his left palate, but it did not cause him any acute pain or difficulty in swallowing.

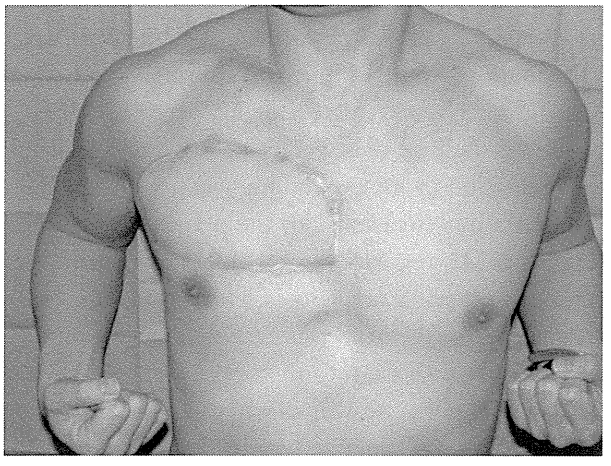

Figure 4. Reconstruction with a latissimus dorsi myocutaneous flap measuring 6 × 18 cm. At 36 postoperative months, the patient's arm movement and activities of daily living were restricted by the latissimus dorsi flap.

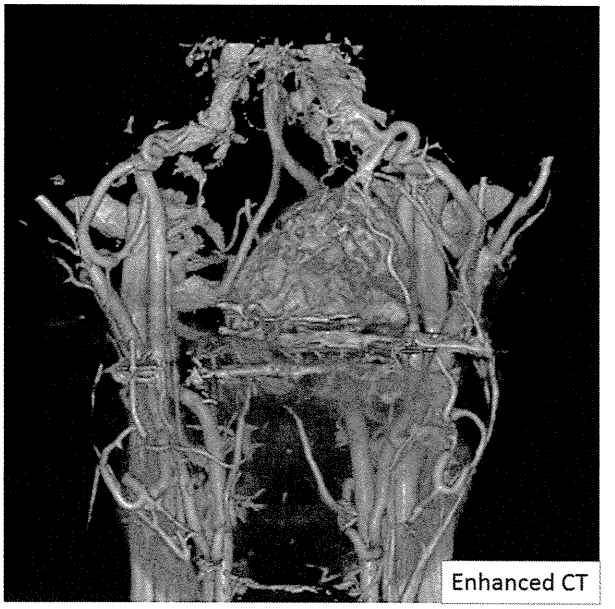

Figure 6. Multidetector CT demonstrated a vascular-rich lesion extending from the upper to middle mucous membranes of the larynx.